Guillain-Barre Syndrome

May 2003

1) THE DISEASE AND ITS EPIDEMIOLOGY

A. Etiologic Agent

The cause of Guillain-Barre syndrome (GBS) is unknown but it is thought to be due to an immune response with cross reactivity to myelin after exposure to an infectious or toxic agent. Infectious agents associated with GBS include different viruses and bacteria. Among bacteria, infections with *Campylobacter jejuni* are found to precede GBS in some cases.

B. Clinical Description and Laboratory Diagnosis

GBS frequently begins with sensory symptoms such as paresthesias (75% of the cases) followed by symmetric flaccid paralysis. Paralysis begins more often in the feet (60%) than hands (20%). Facial and oropharyngeal muscle weakness occurs in up to 60% of cases. Overall 30% of cases will present with respiratory failure.

Electrophysiologic studies are abnormal in 95-99% of patients; these studies show features of demyelination and/or axonal damage and no muscle involvement.

The CBC and sedimentation rate (ESR) are normal in most patients. The cerebrospinal fluid analysis shows increased protein (>55mg/dL in 90% of cases), and the majority of patients will have no CSF pleocytosis (<50 WBC/microliter).

C. Reservoirs

Reservoirs are unknown.

D. Modes of Transmission

Modes of transmission are unknown.

E. Incubation Period

GBS can develop during the 2 months following a symptomatic episode of *C. jejuni* gastrointestinal infection.

F. Period of Communicability or Infectious Period

The disease is not known to be communicable from person-to-person.

G. Epidemiology

Guillain-Barre syndrome occurs worldwide. The annual incidence in the United States is close to 1.0 per 100,000 with the attack rate statistically higher in males (0.52 per 100,000) than in females (0.40 per 100,000). GBS among children less than 15 years old represents more than half of all acute flaccid paralysis cases. In one study, 41% of the severely affected GBS patients showed serologic evidence for infection with *C. jejuni*, cytomegalovirus, Epstein-Barr virus, or *Mycoplasma pneumoniae*, but only 16% of the mildly affected group showed such evidence. Studies from Sweden are showing that the risk of developing GBS during the 2 months following a symptomatic episode of *C. jejuni* infection is approximately 100 times higher than the risk in the general population. Other studies demonstrated seasonal preponderance of GBS in the spring. A possible link between GBS and West Nile virus infection has been examined, and GBS is a criterion for WNV testing during the mosquito season.

2) REPORTING CRITERIA AND LABORATORY TESTING SERVICES

A. New Jersey Department of Health and Senior Services (NJDHSS) Case Definition

CASE CLASSIFICATION

A. CONFIRMED

A clinically diagnosed case supported by:

- Electrophysiologic studies showing nerve conduction slowing with features of demyelination and /or axonal damage, **AND**
- CSF with elevated protein after 1 week of symptoms, and fewer than 50 WBC/microliter, **AND**
- Normal CBC and sedimentation rate (ESR) in most patients.

B. PROBABLE

A clinically diagnosed case not supported by electrophysiological studies.

C. POSSIBLE

Not used.

Note: See Section 3C below for information on how to report a case.

B. Laboratory Testing Services Available

The laboratory features of GBS are nonspecific and nondiagnostic. No testing services are offered at the Public Health and Environmental Laboratories.

3) DISEASE REPORTING AND CASE INVESTIGATION

A. Purpose of Surveillance and Reporting

To identify disease clusters and demographic characteristics.

B. Laboratory and Healthcare Provider Reporting Requirements

The New Jersey Administrative Code (N.J.A.C. 8:57-1.8) stipulates that healthcare providers report (by telephone, confidential fax or in writing) all cases of GBS to the local health officer having jurisdiction over the locality in which the patient lives.

C. Health Officer Reporting and Follow-Up Responsibilities.

1. **Reporting Requirements**

The New Jersey Administrative Code (N.J.A.C. 8:57-1.8) stipulates that each local health officer must report the occurrence of any case of GBS, as defined by the reporting criteria in Section 2 A above. Current requirements are that cases be reported to the NJDHSS Infectious and Zoonotic Diseases Program using the Guillain-Barre Syndrome Reporting Form. A report may also be filed electronically over the Internet using the confidential and secure Communicable Disease Reporting System (CDRS).

2. **Case Investigation**

   a. It is the local health officer responsibility to investigate a case and complete a Guillain-Barre Syndrome Reporting Form. When using reporting electronically, enter collected clinical information into the "Comments" section. Much of the information required on the forms can be obtained from the patient's healthcare provider or the medical record.

b. Use the following guidelines in completing the report:
   1) Accurately record the demographic information, date of symptom onset, whether hospitalized (and associated dates), outcome of disease, and whether the patient has any preceded infection (viral or bacterial, ask specifically about campylobacteriosis), vaccination or surgery in the past 2 months. Collect information about clinical symptoms: muscular weakness, sensory loss. Ask the healthcare provider to submit a copy of the medical record or enlist his/her aid in completing these sections of the Guillain-Barre Syndrome Reporting Form.
   2) Collect as much information as possible about electrophysiologic studies and complications associated with the illness.
   3) Collect information about laboratory tests: cerebrospinal fluid (CSF) examination, erythrocytes sedimentation rate (ESR), cell blood count (CBC).

NOTE: If CDRS is used to report, enter collected clinical and additional laboratory information into the "Comments" section.

   4) If there have been several attempts to obtain patient information (*e.g.,* the patient or healthcare provider does not return calls or respond to a letter, or the patient refuses to divulge information or is too ill to be interviewed), please fill out the form with as much information as possible. Please note on the form the reason why it could not be filled out completely

After completing the case report form, it should be mailed (in an envelope marked "Confidential") to the NJDHSS Infectious and Zoonotic Diseases Program, or the report can be filed electronically over the Internet using the confidential and secure Communicable Disease Reporting System (CDRS). The mailing address is:

NJDHSS
Division of Epidemiology, Environmental and Occupational Health
Infectious and Zoonotic Diseases Program
P.O.Box 369
Trenton, NJ 08625-0369

4) CONTROLLING FURTHER SPREAD

A. Isolation and Quarantine Requirements (N.J.A.C. 8:57-1.10)

None.

B. Protection of Contacts of a Case

None.

C. Managing Special Situations

If multiple cases of GBS occur in city/town, or if an outbreak is suspected investigate clustered cases. Identify common factors, such age, school, workplace or activities to help elucidate risk factors. Consult with the NJDHSS Infectious and Zoonotic Disease Program, at 609.588.7500. The Program staff can help determine a course of action to prevent further cases and can perform surveillance for cases that may cross several jurisdictions and therefore be difficult to identify at a local level.

D. Preventive Measures

None.

ADDITIONAL INFORMATION

A *Guillain-Barre Syndrome Fact Sheet* can be obtained at the NJDHSS website at <http://www.state.nj.us/health>.

There is no formal Centers for Disease Control and Prevention (CDC) surveillance case definition for GBS. CDC case definitions are used by state health departments and CDC to maintain uniform standards for national reporting. For reporting a case to the NJDHSS, always refer to the criteria in Section 2A.

REFERENCES

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